SHORT CASES IN CARDIOLOGY

Supravalvar aortic stenosis: unexpected findings at surgery

C Grahame-Clarke, W B Pugsley, R H Swanton

A 51 year old man was referred for investigation of presumed valvar aortic stenosis. A murmur had been noted as a child but he had developed normally and was entirely asymptomatic. He had had two episodes of infective endocarditis successfully treated medically in 1968 and 1977. He was currently on no medication. Annual echocardiography had shown a possible bicuspid aortic valve with a pressure gradient of 30 mm Hg and a hypertrophied left ventricle with good systolic function.

Examination revealed a fit man with a blood pressure of 140/90 mm Hg in the right arm but only 110 mm Hg systolic in the left, suggesting a preferential streaming up the innominate artery. His pulse was regular at 68 beats/min, which was slow rising in quality. His jugular venous pressure was low and his apex beat was not displaced. He had a long ejection systolic murmur heard all over the precordium radiating to the carotids with no ejection sound.

Electrocardiography showed sinus rhythm with T wave inversion inferolaterally indicative of left ventricular hypertrophy. Chest radiography showed a normal cardiothoracic ratio and aortic arch.

Cardiac catheterisation revealed typical pressure tracings of supravalvar aortic stenosis with a pressure gradient of 50 mm Hg (fig 1). There were giant patent coronaries (fig 2), a dilated innominate artery, and a stenosis at the origin of the left common carotid artery. Right sided pressures were normal with no pulmonary artery aneurysms on pulmonary angiography.

At surgery the aorta was found to be stenotic just above the coronary ostia, which were greatly enlarged. The aortic valve, although tricuspid in architecture, was friable and regurgitant. It was excised and replaced with a 25 mm St Jude's prosthesis. The aortic root wall was unexpectedly rigid, being up to 0.5 cm thick, which caused problems when enlarging the aortic root. Three separate pieces of pericardium were used to construct a gusset, which enlarged the root orifice from 3 mm to 1 cm in diameter. The patient made a satisfactory recovery and was fit and well at one year follow up. Histology of the aortic wall showed medial necrosis with focal calcification and disruption of the elastic lamina in keeping with a diagnosis of atheroma.

Department of Cardiology, The Middlesex Hospital, Mortimer Street, London W1N 8AA, UK
C Grahame-Clarke
R H Swanton

Holmes-Sellors Cardiothoracic Unit, The Middlesex Hospital
W B Pugsley

Correspondence to:
Dr C Grahame-Clarke, The Crucifom Project, The Institute of Neurology, 1 Wakefield Street, London WC1N 1PJ, UK.

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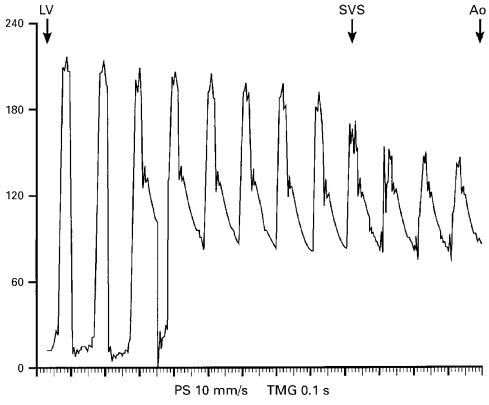

Figure 1 Pressure tracings on pullback of a single lumen catheter from the left ventricle (LV) to the aorta (Ao) showing the supravalvar stenosis (SVS) of 50 mm Hg.

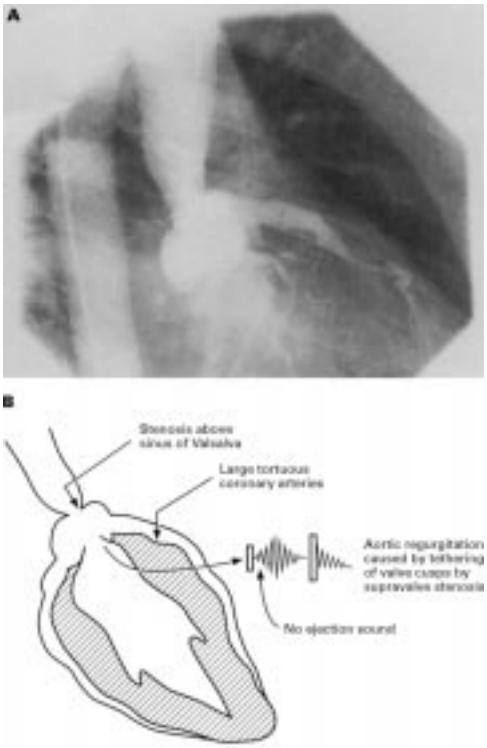

Figure 2 (A) Angiogram from this case and (B) diagrammatic summary of supravalvar aortic stenosis. Reproduced from Pocket Consultant to Cardiology, 2nd ed, 1989, with kind permission of Blackwell Science Ltd.

Discussion

Our patient had no features of Williams's syndrome (mental deficiency, dysmorphic facies, and idiopathic hypercalcaemia), which is known to be associated with supravalvar aortic stenosis. This form of isolated supravalvar aortic stenosis is well known and has previously followed up as valvar aortic stenosis.[1] Multiplane transoesophageal echocardiography has advantages over transthoracic echocardiography and may have helped in our diagnosis.[2] Both isolated supravalvar aortic stenosis and Williams's syndrome are caused by mutations involving the elastin gene on chromosome 7q11.23.[3] Elastin is present in all arterial walls, therefore, it is not surprising that other arteries are affected, as in our case. Although stenoses of the great vessels are rare, dysplasia of the pulmonary, brachiocephalic, and coronary arteries have been reported and can lead to premature coronary artery disease and death even in children.[4] Coronary disease is thought to be exacerbated by high pressure in the coronary arteries secondary to the stenosis. Associated aortic valve disease is strongly correlated to late death and need for reoperation.[5] Therefore, both right and left sided catheterisation are advisable, and early surgical intervention thought appropriate even when the individual is asymptomatic as in our case. It should be remembered that the aortic wall may be thick and difficult to close with a pericardial gusset.

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Sinus bradycardia and multiple episodes of sinus arrest following administration of ibutilide

N B Amin, S Borzak, S Housholder, J E Tisdale

Department of Pharmacy Services, Henry Ford Hospital, 2799 West Grand Boulevard, Detroit, Michigan 48202, USA
N B Amin

Division of Cardiovascular Medicine, Henry Ford Heart and Vascular Institute, Henry Ford Hospital
S Borzak

Department of Nursing, Henry Ford Hospital
S Housholder

Department of Pharmacy Practice, College of Pharmacy and Allied Health Professions, Wayne State University, 230 Shapero Hall, Detroit, MI 48202, USA
J E Tisdale

Correspondence to:
Dr Tisdale.

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Ibutilide is a Vaughan-Williams class III antiarrhythmic agent used for conversion of recent onset atrial flutter and fibrillation.[1] We present the first case report of multiple sinus arrest episodes following administration of ibutilide in a patient with latent sinus node dysfunction.

A 79 year old woman with a history of hypertension, angina, and heart failure presented to the emergency department with exacerbation of heart failure. Medications before admission included metoprolol, digoxin, nitroglycerin, and aspirin.

On day 6, she complained of pleuritic pain radiating to the left arm and neck. Electrocardiography revealed atrial fibrillation with intermittent periods of atrial flutter. She became tachypnoeic with blood pressure of 100/52 mm Hg and heart rate of 82 beats/min. Ibutilide (1 mg) was administered intravenously over 10 minutes for conversion of atrial fibrillation to sinus rhythm, which occurred 10 minutes after infusion (52 beats/min). The patient immediately complained of hot flushes. Over the subsequent 17 minutes she experienced 19 distinct episodes of sinus arrest (mean duration 3.1 seconds, range 2.2–4.5) (fig 1). During these episodes, blood pressure ranged from 103–88/65–51 mm Hg, and heart rate varied from 49–58 beats/min. No treatment was initiated. She remained in sinus rhythm without further sinus arrest episodes during hospitalisation. She experienced no sequelae and was discharged two days later on metoprolol, digoxin, frusemide, and aspirin. She was readmitted three months later with atrial fibrillation (heart rate 84 beats/min) that terminated spontaneously, immediately after which a single 3.3 second sinus pause occurred followed by a junctional beat and then sinus rhythm. Sinus node dysfunction was diagnosed and she had a permanent pacemaker implanted.

Discussion

Single, brief episodes of sinus arrest have been reported following electrical cardioversion of atrial fibrillation.[2 3] Our patient, however, had multiple sinus arrest episodes, which are unlikely to be attributable solely to cardioversion. Ibutilide rechallenge could not be performed because of ethical reasons. Nevertheless, ibutilide was considered a causative or facilitating agent for several reasons. There was a clear temporal relation between ibutilide administration and onset of sinus arrest episodes. Sinus arrest did not occur before ibutilide and did not recur during hospitalisation. Furthermore, the patient had previously experienced two episodes of spontaneous conversion without complications. She had 19 distinct episodes of sinus arrest following ibutilide, compared to only a single sinus pause following spontaneous conversion

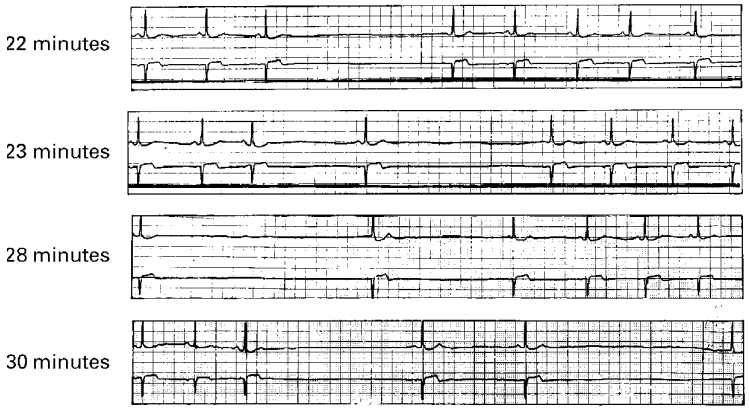

Figure 1 Multiple episodes of sinus arrest following the administration of intravenous ibutilide for termination of atrial fibrillation. On each rhythm strip, the top tracing is lead II and the bottom is lead V1. Times are from start of ibutilide infusion.

during her subsequent admission. Although metoprolol may inhibit sinus node function, she had been taking this drug for five months before admission without complications. She was on a stable dose of digoxin for one month with a therapeutic serum digoxin concentration. Myocardial infarction was ruled out.

Ibutilide in the presence of metoprolol and digoxin likely unmasked and exacerbated underlying sinus node dysfunction in our patient, resulting in multiple episodes of sinus arrest. Ibutilide should be used cautiously in patients with documented or suspected sinus node dysfunction.

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Azienda Ospedaliera S Carlo, Invasive Cardiology, I-85100 Potenza, Italy
W Serino

Royal Brompton Hospital, Sydney Street, London SW3 6NP, UK
U Sigwart

Correspondence to:
U Sigwart.

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Septal ablation in a patient with hypertrophic obstructive cardiomyopathy and a unique variant of anomalous origin of the left coronary artery

W Serino, U Sigwart

A 71 year old woman was referred for assessment of a severe form of hypertrophic obstructive cardiomyopathy. She complained of chest pain and shortness of breath on exertion. She had an uncertain family history of heart disease, but there was no family history of sudden death. Her history included hypothyroidism, hypertension, and cerebrovascular accidents with minimal sequelae. She had moderate stenosis of the right internal carotid artery. The patient was taking verapamil (240 mg/day) and atenolol (50 mg/day).

Physical examination revealed a 3/6 systolic ejection murmur radiating towards the neck; there was no ejection click. Blood pressure was 180/100 mm Hg and she was in sinus rhythm (heart rate 57 beats/min).

Electrocardiography showed left ventricular hypertrophy with giant T wave inversion and ST segment depression in the anterior and lateral leads. Chest radiography showed an increased cardiothoracic ratio of 16:27. Cross sectional echocardiography showed a 3.8 mm thick intraventricular septum. The left ventricular outflow tract gradient was 65 mm Hg at rest and increased to 120 mm Hg with dobutamine infusion. There was mild to moderate mitral incompetence.

The patient underwent left and right heart catheterisation, and a transeptal approach was used for left ventricular pressure monitoring. Mean outflow tract gradient at rest was 35 mm Hg, which increased to 150 mm Hg after 2 mg of dobutamine (fig 1A).

Coronary arteriography showed a relatively small left coronary artery giving branches

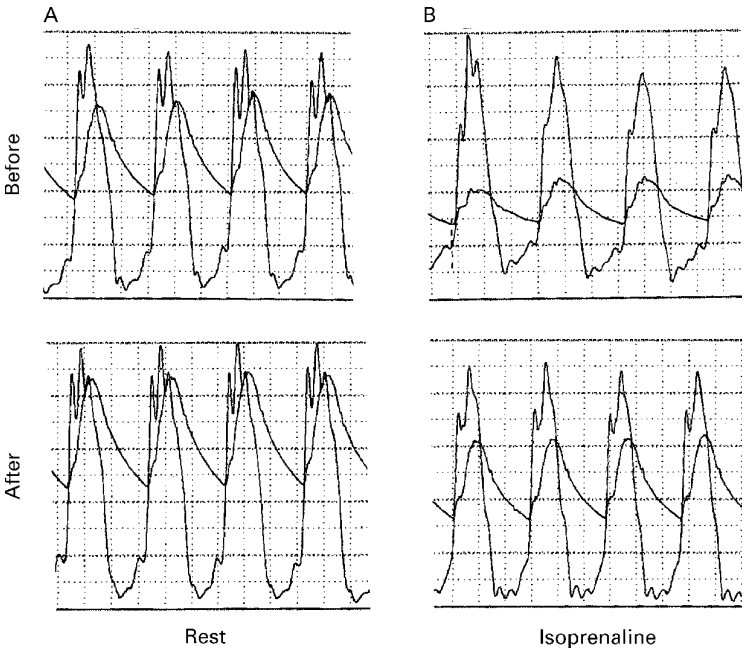

Figure 1 Simultaneous left ventricular and aortic pressure tracings at rest and after provocation with intravenous isoprenaline. The resting left ventricular outflow tract pressure gradient is completely abolished after the procedure, the very high pressure gradient after isoprenaline provocation is largely reduced following the procedure.

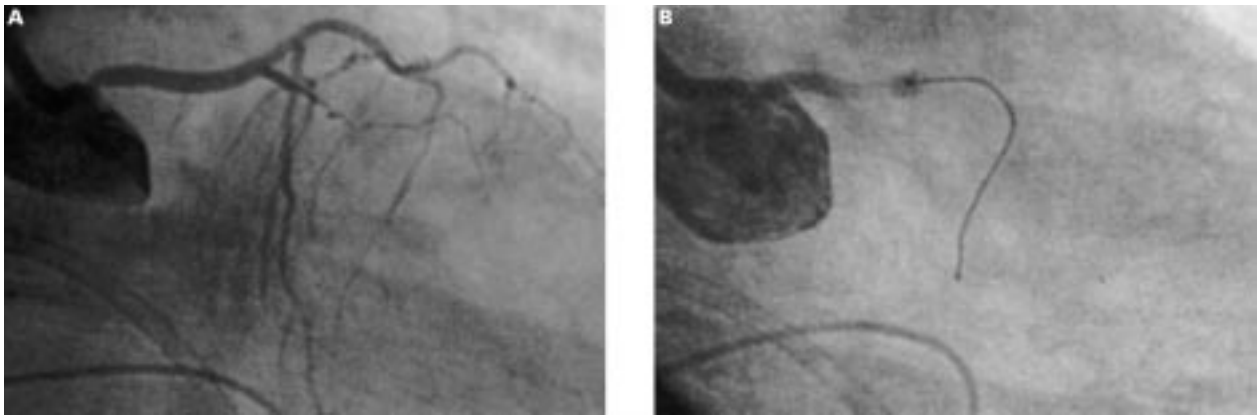

Figure 2 Angiographic documentation of the "left" coronary artery (A) showing a number of septal perforators originating from one common artery, which has its origin in the left sinus of Valsalva. (B) Presence of an over the wire balloon catheter in the principal artery. The balloon obstructs the lumen completely. The angioplasty guidewire is seen in the largest septal perforator. The transeptal Brochenbrough catheter and the pacemaker electrode in the right ventricle are seen in the left lower corner.

directed vertically into the intraventricular septum (fig 2A). Selective right coronary arteriography showed a dominant right coronary artery with the circumflex arising from the proximal part of the right and running posteriorly between the aorta and the pulmonary artery. The left anterior descending coronary artery originated from what appeared to be a large conal branch, putting this anomaly into the extremely rare group of single coronary artery of the right "malignant" type. The persistence of the solitary left septal perforator makes this a unique finding. All three major branches were free from degenerative changes.

Alcohol ablation[1] of the first small septal branch of the isolated left coronary perforator was carried out but with no significant effect on the resting gradient. A balloon was then inflated in the main trunk of the "left" coronary artery about 2 cm away from the ostium (fig 2B). During temporary occlusion of this branch, the systolic gradient disappeared and even isoprenaline provocation failed to provoke any significant pressure gradient. It was therefore decided to ablate the entire "left" coronary artery, which appeared to be responsible for most of the upper septal perfusion. This was done by an injection of 5 ml of absolute alcohol during which the patient complained of serious chest pain. There were more than 3 mm anterolateral and inferior ST segment depression and some extrasystoles often in couplets. At the end of the procedure the resting gradient was abolished and the peak gradient during isoprenaline stress was 45 mm Hg (fig 1B).

There was a moderate rise in creatine kinase (CK) (2220 U/l; CK-MB 269 U/l) and ECG changes reverted to normal in a few hours. Cross sectional echocardiography performed 24 hours later showed a resting gradient of 35 mm Hg and a reduction of the septal thickness to 2.7 mm Hg. Mitral valve incompetence was unchanged. The patient reported profound clinical improvement at six weeks' and seven months' follow up.

Therapeutic occlusion of this, probably unique, separately originating septal system resulted in dramatic haemodynamic improvement. The risk of suppressing this vessel seemed minimal in view of the excellent state of the right coronary artery that supplied the rest of the myocardium.

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